



## Kleine-Levin Syndrome- a Syndrome of Periodic Hyper-Somnia: a Case Report

### KEYWORDS

Kleine-Levin syndrome, hypersomnia, derealisation, lithium

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**ABSTRACT** *Kleine-Levin syndrome (KLS) is an uncommon syndrome characterized by recurrent episodes of hypersomnia, behavioural and/or cognitive disturbances, abnormal eating and hypersexual behaviour. The diagnosis is clinical and requires a high index of suspicion and exclusion of other neurological conditions and sleep disorders. We report a case of 17 year old girl who had recurrent episodes of hypersomnia with derealisation, suicidal ideations and food fads; and a good response to lithium therapy.*

### Introduction:

Kleine-Levin syndrome (KLS) or Rip van Winkle syndrome was defined by Critchley as, "a syndrome composed of recurring episodes of undue sleepiness lasting some days, associated with an inordinate intake of food, and often with abnormal behaviour".<sup>[1]</sup> It is an uncommon symptom complex characterized by recurrent episodes of hypersomnia, behavioural and/or cognitive disturbances, abnormal eating and hypersexual behaviour.<sup>[2,3]</sup> The disease is of uncertain aetiology and predominantly affects adolescent males. It is a benign disorder, shows complete spontaneous remission without any neurological sequelae and a good response to the treatment.<sup>[2,3]</sup> It is often underrecognized and mistreated as a psychiatric disorder. We report a case of Kleine-Levin syndrome in an adolescent girl with good response to lithium.

### Case Report:

A 17 year old schoolgirl presented with a two year history of recurrent episodes of excessive sleepiness. The episodes would be preceded by a feeling of marked fatigue which would gradually culminate into an irresistible desire to sleep. For the initial two days, she would sleep nearly 20 hours a day. In her waking hours, she would be irritable, refuse to eat, would cry, sing sad songs and have vivid suicidal ideas. Over the following days, the hours of sleep would wane off and her appetite would increase abnormally. She would refuse the usual foods and demand spicy potato chips and cold drinks. Feeling as if in a dream world, she would insist on her mother's company at all times. The episodes would usually last for 8-10 days and remit abruptly. In between the episodes, she would remain normal.

Her first episode occurred two years previously, 15 days after a febrile illness. The initial frequency was once every two to three months, which gradually increased to twice a month. She denied use of any medications or any substances.

Two of her episodes were witnessed, wherein her excessive sleepiness and food fads were noted. The most troublesome was her feeling of derealisation, during which she would doubt whether her family members and attending doctors were actually real. Both the episodes terminated abruptly. She was however, able to remember all the events that occurred during the episodes. A provisional diagnosis of periodic hypersomnia was entertained.

Her general, systemic and neurological examination was unremarkable. All biochemical investigations including thyroid functions and serum ammonia levels were normal. Electroencephalography (EEG) showed intermittent generalised 5-7 Hz theta waves during the episodes and was normal between attacks. Polysomnography (PSG) showed normal sleep architecture. Brain MRI was normal. A final diagnosis of Kleine-Levin syndrome was made.

She was started on armodafinil 150 mg/day on which her somnolence decreased, but the derealization during the wakeful period worsened distressingly. Armodafinil was stopped. Lithium carbonate was started at a dose of 600 mg/day, after which she has had no further episodes in the past two years of follow-up.

### Discussion:

Our patient fulfilled the International Classification of Sleep Disorders II criteria for KLS, which include<sup>[4]</sup>:

- A) Recurrent episodes of severe hypersomnia (2-31 days)
- B) One or more of the associated features:
  1. Cognitive abnormalities such as feeling of unreality, confusion, hallucinations,
  2. Abnormal behavior such as irritability, aggression, odd behaviour,
  3. Binge eating,
  4. Hyper-sexuality
- C) Interspersed with long periods of normal sleep, cognition, behavior and mood
- D) The hypersomnia is not better explained by another sleep disorder, medical, neurological or mental disorder, medication use, or substance use disorder.

In the largest systematic review of 186 cases of KLS, the mean age of presentation was 17 years, and a male predominance (Male: Female- 2:1) was noted. The onset is frequently associated with a precipitating factor, most commonly an infection, as in our case. Other precipitating factors reported are alcohol, sleep deprivation, head trauma, physical exertion, mental stress and drug abuse.<sup>[2,3]</sup> Hypersomnia is the universal feature (100%), ranging between 12-24 hours/day.<sup>[1]</sup> Behavioural disturbances including derealisation, confusion, abnormal speech, hallucinations, delusions, depressed mood, irritability and aggressive behaviour have been re-

ported in 96% cases.<sup>3</sup> However the feeling of unreality or disconnected thinking during the episodes, as was prominent in our case, was reported to be the most specific symptom of this syndrome.<sup>[3]</sup> Arnulf et al reported hyperphagia in 78% cases and hypersexual behaviour in 43% cases. Apart from hyperphagia, the patient may have craving for special foods, excessive drinking, or aversion to food. Patients may or may not retain memories of events that occurred during the episode, and most of the patients remain normal in the inter-episodic periods<sup>[3]</sup>.

The most striking feature in our patient was the feeling of derealisation during the episodes. There was no hypersexual behaviour and memory for events during the episodes was retained.

In about 75% cases, EEG demonstrates diffuse slowing of the background during the episodes. Less often, EEG may show low frequency, high amplitude delta or theta waves predominantly in temporal or frontotemporal regions, occurring in isolation or in sequences.<sup>[2,3]</sup> Rarely, isolated spikes or sharp waves may be seen.<sup>[5,6]</sup> In our case EEG showed intermittent generalised 5-7 Hz theta waves during the episodes and was normal in between attacks.

The pathogenic basis of KLS remains an enigma. However, functional neuroimaging has suggested the possible role of hypothalamic, thalamic and frontotemporal dysfunction.<sup>[7,8]</sup> Secondary KLS has been described in the setting of traumatic brain injury, stroke, infections, autoimmune and paraneoplastic encephalitis.<sup>[2,3]</sup> An association with HLA-DQ2 has been proposed in view of the familial clustering and the association with autoimmune disorders.<sup>[9]</sup> HLA typing could not be done in our patient.

KLS is a benign disorder, and in most patients the episodes wane off spontaneously with time, within a median duration of 8 years.<sup>[3]</sup> There is no consensus on the treatment of choice for KLS, however amphetamines, amantadine and modafinil have been reported to improve the hypersomnolence.<sup>[2,3]</sup> However, the alleviation of somnolence may unmask more distressing symptoms like derealisation as was dramatically evident in our case. Lithium has been found to reduce the frequency of the episodes.<sup>[2,3]</sup> Our patient also showed a good response to lithium.

We conclude that Kleine-Levin syndrome is an uncommon benign treatable disorder, which requires a high index of clinical suspicion.

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